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Tubular duplication of colon and terminal ileum in a female child, case report, review of literature and proposal of a new classification

Mukul Kothari*

Maternity and Children's Hospital, Al Morikabat, P.O. Box 2595, Dammam 31176, Saudi Arabia

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ABSTRACT

A case of a four and a half years old girl with total colon and terminal ileal duplication with a normally situated anus, vestibular fistula, double bladder and urethra with a unique feature of loop duplication of terminal ileum and part of the colon is reported. A proposal is made for a new simplified classification.

Alimentary tract duplications are unusual malformations with an incidence of 1:4000–5000 [1]. Incidence of colonic duplications vary from 4 to 30% in various series [2–4]. Even rarer are the tubular duplications that affect several segments of the colon or the entire colon. A case of a four and a half year old girl who presented with fecal discharge from a vestibular opening in addition to normally situated anus is reported. She was found to have a complete duplication of colon and terminal ileum ending in the vestibular fistula. This had unique features of completely separate blood supply to part of the duplication and associated complete duplication of urethra and bladder. Varying terms have been used in the literature to describe duplications affecting these segments of bowel so we propose use of unified terminology in describing such cases. A simplified classification is also suggested to include all varieties of total colon and hindgut duplication.

1. Case report

A four and a half years old Sudanese girl was brought to the clinic by her parents with the complaint of fecal discharge from a small opening in the vestibule. This was noticed soon after birth. She was a product of full term normal vaginal delivery. Her parents are not related. There was no history of constipation, abdominal pain or urinary problem. She had regular bowel movements through normally situated anus. They were advised to wait until later age for the treatment. She underwent a contrast study through normally situated anal opening in Sudan at the age of 4 years which showed no abnormality or a fistulous connection to the vestibule. On examination she had normal looking external genitalia with a normal anus and an opening in the vestibule just behind the vaginal opening discharging feces. There were no other obvious associated anomalies. She was planned for an examination under anesthesia (EUA). What intrigued us was the observation that the fistula continued to discharge feces even when the washouts from the rectum were clear after two days of preparation.

EUA revealed presence of a duplicated bowel with a narrow fistula opening that would allow only size 10 feeding tube. Later she underwent simultaneous contrast study through normal anus and the fistula opening which showed a duplication in front of normal rectum extending right up to the sigmoid and then apparently

* Corresponding author. Tel.: +966 533213848.
E-mail address: mukulkothari@gmail.com.

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communicating with the sigmoid colon (Fig. 1a). The radiologist terminated the study at this point thinking that this is a tubular duplication with proximal communication at sigmoid level. CT with contrast to look for associated anomalies showed that the spine was normal but the bladder was found to be completely duplicated with normal kidneys with each side draining in to ipsilateral half of bladder (Fig. 1b).

At the time of definitive surgery closer examination of genitals and cystourethroscopy with vaginoscopy showed that she had double urethra and two separate bladders. There was a high vertical vaginal septum with separate cervices on each side. Abdominal exploration revealed that duplication involved whole of the colon and terminal 20 cm of ileum (Fig. 2a). There were two appendix and two cecum (Fig. 3a). The colonic duplication was on the mesenteric side. Terminal 20 cm of ileum, ascending colon and half of transverse colon had a separate mesenteric supply (Looped duplication). Beyond this the duplicated colon was fused to the native colon in a double barrel fashion. There was a thick mesentery like structure at the common wall right up to recto-sigmoid junction (Fig. 3b). Mesentery of the looped duplication was fenestrated and the blood supply was from a single vessel coming off from a terminal ileal vessel (Fig. 3c). Proximally the duplication joined the normal ileum in Y-shape fashion. The fistulous opening was dissected submucosally from perineum. The dissection was carried out to the level of peritoneal reflection. Abdomen was entered through a hockey-stick incision extending to the left. The duplication was opened about 5 cm above the peritoneal reflection and submucosal dissection was done to core out mucosal tube which was dissected from below. The muscular cuff was left in place. The distal end of the remaining duplication was anastomosed end to side to the recto-sigmoid junction of the native colon after division of common wall proximally with a stapler for about 10 cm. Parts of the duplication with separate blood supply including ileum, cecum with appendix, ascending colon and the mid transverse colon until the point where it fused with the native colon were removed completely. The muscular spur at the proximal end of the duplication where it joined the native ileum was removed with the stapler and the ileum was closed with two layers of interrupted sutures. The opened duplication at the level of mid transverse colon was closed over completely with two layers of Vicryl sutures. This left a tubular duplication in place with proximally closed end with anastomosis at the level of recto-sigmoid into the native colon (Fig. 2b). A proximal transverse colostomy was brought out through the umbilicus. The urinary tract was left untouched. She made an uneventful post-operative recovery and was started on diet after the colostomy started functioning. The Histopathology of the excised segments of the bowel did not reveal any ectopic mucosa. A contrast study before the closure of the colostomy (Fig. 4) showed no reflux in to the remaining duplication. At one year follow up she continues to do well with no constipation or bowel related complaints. She continues to pass urine normally every 3 to 4 hour.

2. Discussion

Such rare are the duplications affecting entire colon that Yousefzadeh et al. [5] encountered only 54 cases described in the English language literature from the time first case was described by Suppiger in 1876 [6] until the publication of their report in 1983. They added 3 new cases taking the total to 57. Bower et al. [7] reported 3 cases of total colonic duplication in their experience of 78 cases of alimentary tract duplications over 40 years. Macpherson in 1993 [3] analyzed four large series with combined 281 cases of gastrointestinal tract duplications. In his review colorectal duplications accounted for 6% of all gastrointestinal tract duplications. Except for the three large review studies published on the condition [3,5,6] total colon and hindgut duplications reported in medical literature are usually isolated case reports.

There is lack of uniform terminology in describing duplications affecting colon. Some of the terms used to describe cases of duplications involving entire colon and in some cases, terminal ileum in the literature are, 'colorectal duplication [3],' 'tubular colonic duplication [5],' 'hindgut duplication [6]' and 'total colon duplications [7].' Term 'colon duplication' is also applied to cystic or tubular duplications affecting any segment of colon as well as to those long tubular duplications affecting several segments of colon. Similarly the term 'hindgut duplication' is applied to duplications affecting isolated segments of colon and to those involving terminal ileum. Use of uniform terminology is required for reporting these cases in the literature. We propose that the term *hindgut duplication* should only be applied when the duplication involves the derivative of the embryological hindgut i.e. the bowel beyond distal half of the transverse colon. *Total colon or complete colon duplication* is the term that should be applied to the duplications involving four or more segments of colon without involvement of ileum, while colon and

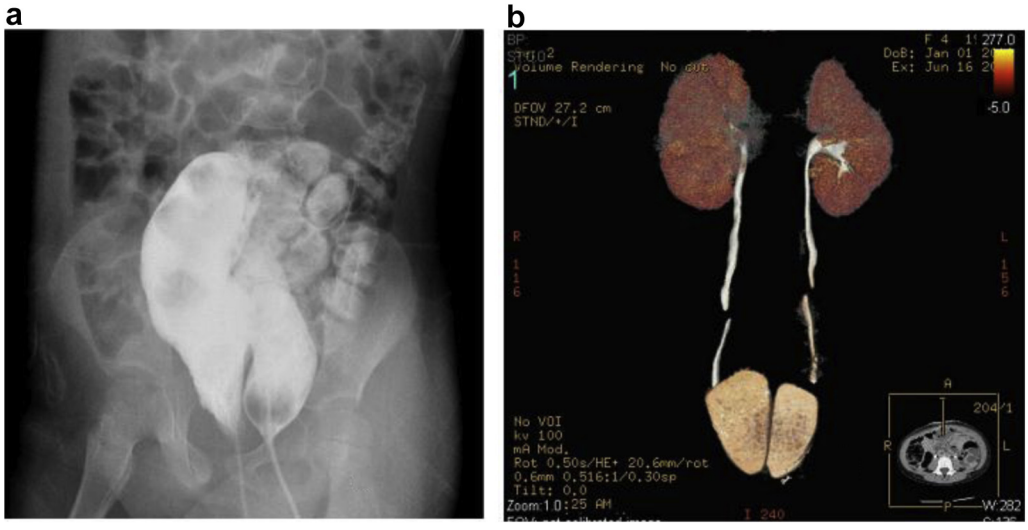

Fig. 1. a) Preoperative contrast showing duplication, b) CT scan showing duplicated bladder and each kidney draining in to ipsilateral bladder.

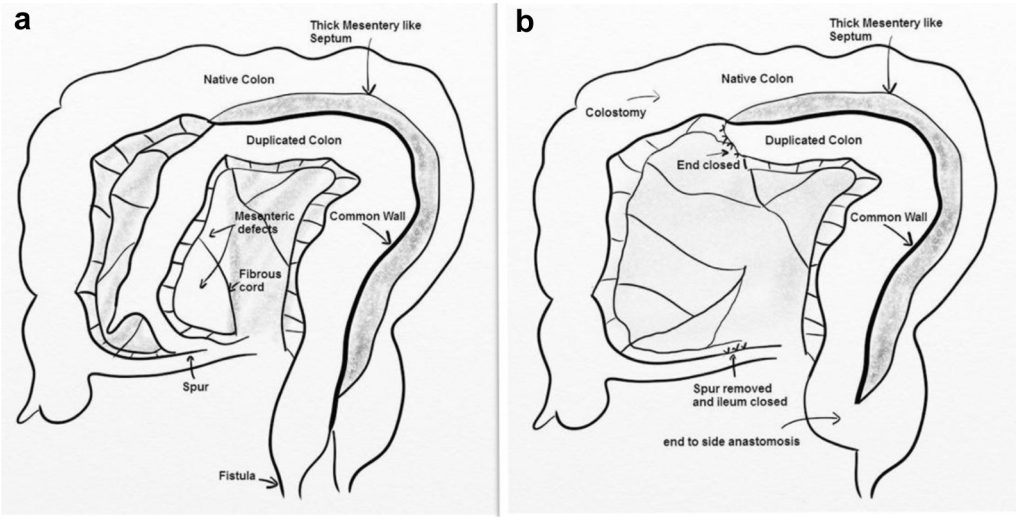

Fig. 2. Drawings detailing findings at laparotomy (a) and final anatomy following the surgical procedure (b).

ileal duplications should be described by the term *tubular duplication of colon and terminal ileum*. The other cystic and segmental tubular duplications should be denoted by the segment involved. This case describes a tubular duplication of terminal ileum and colon.

Yousefzadeh et al. in their review article in 1983 [5] mentioned that no two similar cases have been described until then. Most cases differed in the extent of duplication of the genitourinary tract and type of the associated malformations. With the passage of time few case reports have appeared that describe cases identical to the ones previously described in the literature [4,8,9]. These cases have same anatomy of the duplicated colon without associated malformation. During the search of English language literature we came across only 4 cases of total colon duplication with a normal anus and a vulvar or vestibular fistula [4,6,9] and 6 cases of vaginal fistula [8,10,11] similar to the present case. Most of these differed in the involvement of ileum and other associated anomalies. Our case is unique with complete urethra and bladder duplication along with high vaginal septum and double cervix. Our case also had unique loop duplication as described above.

Loop duplication was the term used by Kottra and Dodds [12] in 1971 describing a duplication with separate mesentery and blood supply. They also noted that until then it was reported only in the small bowel. A recent report by Chooramani et al. in 1997 [13] described a male child with hindgut duplication having double mesentery of the almost entire segment except the distal most rectum allowing them to excise the duplication without affecting the normal bowel. In our case there was loop duplication of terminal ileum and the colon up to the mid transverse segment. The mesentery of this segment was fenestrated, divided by a single fibrous band. The segment was excised without affecting the normal colon. The excision was performed to prevent internal herniation.

Treatment of patients with long tubular colon duplications involves consideration of associated anomalies in addition to the treatment of duplication. Fortunately in most cases with a normally situated anus and a vestibular, vulvar or vaginal fistula of the duplicated colon, associated anomalies are less common and less severe [4,8–11] and hence the treatment of the duplication only is usually sufficient. Various surgical techniques have been described in the literature to deal with the duplication with a fistulous opening like the case described here. Riedel operated on Grohe's case described in 1900 [5]. He divided the septum between two rectums and the tissue between anal orifices and reconstructed the perineum. Brunschwig [14] performed a lateral anastomosis between the two sigmoid and the sigmoid leading to the fistula was divided below the anastomosis leaving an intestinal sac to discharge mucus through the fistula. Similar technique was used in a case described by Sarpel et al. [11] recently in 2005. Yucesan et al. [4] in 1986 dissected the fistula tract from below and created a large anastomosis between the duplication and the normal colon to eliminate any blind end. Kaur et al. [9] created a communicating window of 8 cm between the two sigmoid and the rectum as low as

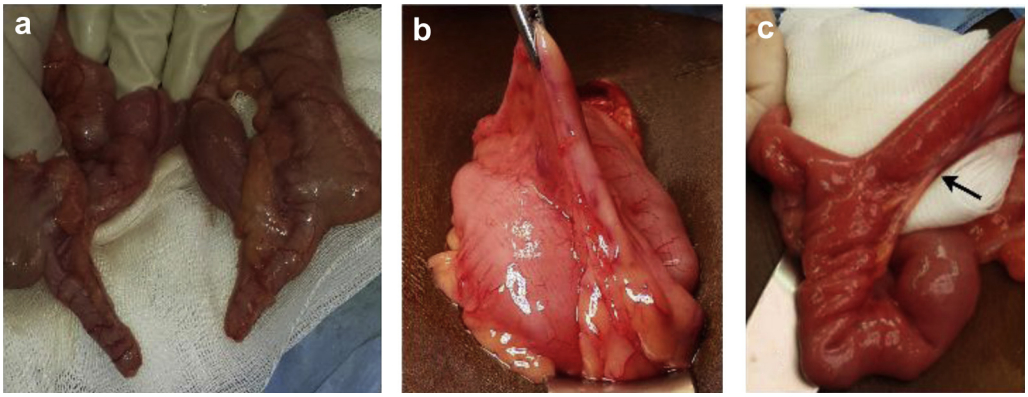

Fig. 3. a) Double cecum and appendix, b) double barreled segment, c) Y shaped proximal communication, arrow points to edge of the defect in mesentery with single artery.

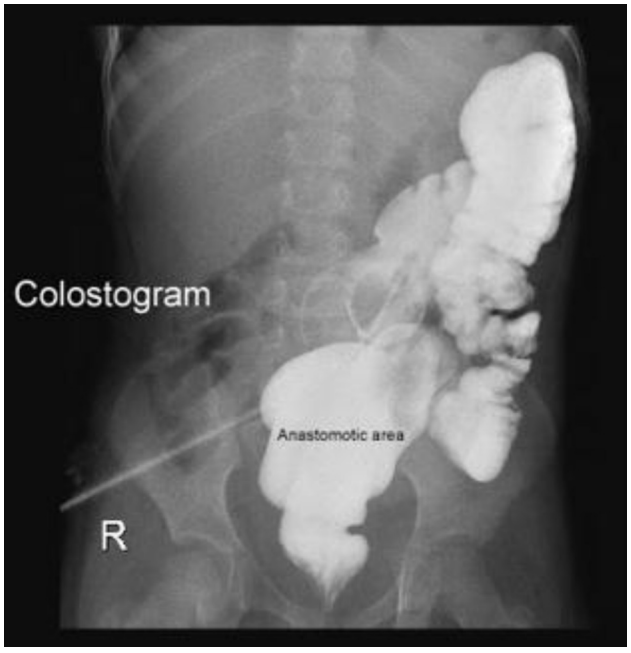

Fig. 4. Colostogram showing anastomotic area without reflux in to remaining duplicated bowel.

possible with excision of fistula from below. Recurrence of rectovaginal fistula was noted in their patient. We dealt with our case on same principals as described by McPherson et al. [8] in 1969 with anastomosis of the sigmoid to its twin and removal of mucous membrane from the lower segment.

Attempts have been made to classify colon duplications in to various categories [5,12]. Yousefzadeh et al. [5] described a modification of Smith's classification [15] based mainly on the existence of fistula(e) describing five groups but again this is not consistent across the groups. The present case belongs to group 2 of their classification. Kottra and Dodds [12] described a classification based on presence of symptoms and associated genital and urinary tract anomalies. Several cases described recently [8–11] with rectovestibular, rectovulvar or rectovaginal fistula without any associated anomaly cannot be classified according to their system. Craigie et al. [16] encountered a male neonate with long Y duplication of colon and imperforate anus with one colon ending in a rectourethral fistula and the other ending blindly. Unable to classify their case with established systems they proposed a practical classification based on status of colon proper and the duplicated colon. According to this scheme there are 9 possible scenarios depending on how each colon ends. We propose a following simple classification (Table 1) based on study of cases reported in the literature. This is based simply on presence or absence of a normal looking anus/ani and how the duplicated bowel ends. It takes care of all possible variations observed in the literature. Most of the authors reporting the colon duplications have not classified the anomaly based on either of the two classifications reported so far and that is because these classifications [5,12] cannot accommodate all the variations. These classifications are also quite complex. The classification described here is simple, self-explanatory and easy to remember and yet points out all possible variations. It also guides one to look for the type of the anatomic arrangement and associated anomalies that can be present. It is important to remember that duplications and other complex anomalies of genitourinary tract, spinal column and cord malformations can be present in any case and should be investigated with contrast studies, CT scan and MRI as necessary. The cases of duplications of colon with absent anus are usually diagnosed surreptitiously during the course of treatment for the imperforate anus. Genitourinary anomalies are more complex and difficult to treat in cases with absent anus. The prognosis of fecal and urinary continence in these cases is guarded and will depend on the anatomy.

Table 1
Classification for Hindgut and total colon duplications (with or without duplication of terminal ileum).[a,b]

Group 1: Anus present (single or double)
- a) Single perineal anus
  1. Duplicated colon ending in a genitourinary (urethral, vaginal or vestibular) fistula
  2. Duplicated colon ending blind
  3. Duplicated colon with distal communication to the native colon
- b) Two perineal ani
  1. In coronal plane
  2. In sagittal plane

Group 2: Absent anus[c]
- a) Both colons end blind
- b) Both colons terminating in a fistula to genitourinary tract
- c) One colon ends blind, one terminating in a fistula to genitourinary tract

[a] Associated duplications of the genitourinary tract, anomalies of spinal column and cord can be present with any of the subcategories and patient should be investigated in detail looking for them. In general those cases with absent anus have more complex associated anomalies and are difficult to treat.
[b] Duplication can be on the mesenteric or antimesenteric side of the native colon.
[c] Cases with absent anus are usually diagnosed surreptitiously and the prognosis with regards to continence will depend on the anatomy.

3. Conclusion

In summary we describe an unusual case of female with total colon and terminal ileum duplication and rectovestibular fistula associated with complete duplication of urethra and bladder with partially duplicated vagina and double cervix. The case was treated on established surgical principals of preservation of colon length, distal end to side anastomosis of the duplicated segment to the native colon ensuring emptying of the segment and eliminating blind end and complete submucosal excision of the fistulous connection preserving integrity of vagina and normal pelvic musculature. We also propose a simple all-encompassing classification of hindgut and total colonic duplication.

Conflicts of interest

None.

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